Original Article

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Management of Conjoined Twins During Neonatal Period

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***Background/Purpose**: Conjoined twins are rare and complex anomalies of the newborn. It is reported in 1/50.000 to 1/100.000 live births. The aim of the study was to summarize the experiences gained during separation of 5 sets of conjoined twins with presentation of literature review.*

***Materials & Methods**: During the period from January 2003 to June 2009, 5 sets of Conjoined twins had been separated during the first month of life. Two sets were symmetrical while the other 3 sets were asymmetrical.*

***Results**: Urgent separation of 3 sets of conjoined twins had been performed due to respiratory embarrassment. Elective surgery was performed for the case presented as fetus in fetu and the case of thoraco- omphalopagus twins in which the shared liver was divided between both babies. The short term postoperative follow up revealed uneventful course of 6 children resulting from separation of 5 sets of conjoined twins.*

***Conclusion**: Parasitic twins and twins with sever anomalies incompatible with life in one of them are considered to be one person. Timing of separation and separation plan should be individualized according to the need of urgent separation and the degree of organ fusion.*

***Index Word**: conjoined twins, management, neonatal period.*

INTRODUCTION

Identical twins are the result of complete separation of the inner cell mass (totipotent cells) at up to 7 days after fertilization, on the other hand, conjoined twins(CT) are probably the result of monozygotic twins in which the embryonic disk divides later causing incomplete fission [1]. The condition is associated with incomplete differentiation of various organ systems [2]. Such twins are identical in sex and karyotype [3].

The incidence of conjoined twins is reported to range from 1 in 50.000 to 1 in 100.000 live births (4) with a male to female ratio of 1 : 3 [5,6].

Diplopagus is the well known form of CT in which both members are equal in size and symmetrically joined to each other (7). The most common variety is the thoraco-omphalopagus type, accounting for 30-40% in most series [8,9] and 75% in other series [10].

In contrast to diplopagus, in heteropagus form, which is extremely rare, one complete member (autosite) bears an incomplete one (parasite) that is dependent on the normal member. The parasite is attached to the autosite in a nonduplicated fashion to any portion of the body or even within the body as fetus-in-fetu [10].

CT are classified into eight basic types with each type posing its own reconstructive challenges [11] not only with the division of shared organs but also with the closure of a large defect [12].

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The first successful separation of CT was performed in 1689 by Johannes Fatio [13], while the most popular CT was Chang and Eng Bunker born in Siam in 1811. They were not separated and married two sisters and had 22 children [14]. The aim of this study was to summarize the experiences gained during management of 5 sets of conjoined twins with special emphasis on the frequency, types and timing of separation of CT presented at (MUCH)

PATIENTS AND METHODS

During the period from January 2003 to June 2009, 5 sets of Conjoined twins had been separated during the first month of life. Two sets were symmetrical while the other 3 sets were asymmetrical.

Case 1

Male epigastric heteropagus conjoined twins were delivered at January, 2003 by C.S in the first pregnancy of a 24 year- old lady, their combined weight was 4.8kg.

The parasitic one was without head and neck, with rudimentary cardiopulmonary system but other anatomical features were normal like the autosite. They were joined from the xiphisternum to the umbilicus. (Fig. 1)

C.T scan revealed that both livers were just touching each other without vascular or biliary connection.

The viscera of both twins were completely separated. The process of separation was successfully carried out at 4th day after delivery as the weight of the parasite on the upper abdomen of the autosite caused respiratory embarrassment. Closure of the defect of the complete member was easy with skin which was taken from the parasitic one.

Post operative period was uneventful.

Case 2

Female pygopagus CT were born on Jun, 2004 by C.S, the third pregnancy of 35 year- old mother, their combined weight was 5.850kg.

Both were born alive, computed tomography and echo cardiography were performed after admission. One of the twins had craniorachischisis with single chamber heart while the other baby had small VSD. The area of sharing was the sacral part of the vertebral column. (Fig. 2)

The baby with craniorachischisis had imperforate anus while the other baby had normally situated one, both neural tubes were in continuity, separation was performed in the 3rd day of life urgently due to death of the malformed twin. The process of separation continued for 3½ hours only and the skin defect on the sacrum of the living baby closed primarily.

Case 3

Female infant presented at birth on December, 2005 with abdominal mass causing respiratory embarrassment.

Abdominal X-ray revealed areas of calcification within the mass. Abdominal C.T revealed a solid mass within the root of mesentery with ossified tissues inside, so teratoma was the preoperative diagnosis.

On exploration, the mass was covered with scalp like tissue containing hair; the mass was excised completely and on histo-pathological examination, 4 vertebral bodies were present in addition to other elements of intestinal, renal and muscular tissues, so the case was diagnosed as fetus-in fetu. (Fig. 3)

Case 4

A female baby with parasitic teratopagus twin protruding from the mouth and causing severe respiratory distress. The baby was delivered by C.S at February, 2005 to a second gravida 34 year- old mother. (Fig.4).

Emergency excision of the parasitic twin was performed after 8 hours of delivery. The parasitic twin root was pedunculated and attached to the hard palate of the normal baby. After excision, intermaxillary, wide defect cleft palate was observed.

Histopathological examination of the excised parasite revealed vertebral bodies, brain tissue occupying the bulky end and semiformed small upper and lower extremities, all tissues were mature so no recurrence occurred.

The cleft palate repair was performed at one year of age.

Case 5

On June 2009, male thoraco-omphalopagus twins were diagnosed antenatally and delivered by C.S at 38 weeks of gestation in the second pregnancy of a 26 year- old lady. Their combined weight was 5.25kg.

Examination revealed that the area of fusion included the xiphisternum and the ventral aspect of 9th and 10th ribs, epigastrium and umbilicus. (Fig.5-A)

Helical computed tomography with I.V contrast revealed that both babies shared a centrally situated liver with the right lobe in the abdomen of one twin while the left lobe was present in the bridge between both twins. Each of both segments of the liver had a separate venous drainage and biliary systems. The I.V dye was injected in one twin and was excreted in the renal system of both twins after about half an hour. Administered oral contrast ensure complete separation of G.I.T of both twins.

Separation was performed at one month of age and continued for 8 hours during which the right and left lobes were separated with the finger fracture technique and electrocautery, with suture ligation of the crossing blood vessels and bile ducts. It is found that each lobe had its own biliary system, and separate inferior vena cava. After separation, skin closure was only feasible after performing lateral release incisions for one of the babies while the other baby's wound was closed easily. (Fig. 5- B,C)

Post operatively, the infant whose wound was closed with skin only was in need for artificial ventilation for 3 days and after that he developed gastro-esophageal reflux that was treated conservatively. (Fig.5-D)

RESULTS

Urgent separation of 3 sets of conjoined twins had been performed due to respiratory embarrassment in cases of epigastric heteropagus twin and the case of oral teratopagus twin. The 3rd process of urgent separation was in case of pygopagus twin due to death of the malformed member – dictive surgery was performed for the case presented as fetus in fetus and the case of thoraco omphalopagus twin in which the shared liver was divided with finger fracture teaching.

The short term post operative postoperative follow up revealed uneventful course of 6 children resulting from separation of 5 sets of conjoined twins.

Fig. 1: Male epigastric heteropagus twin

Fig. 2: Female Pygopagus Twin

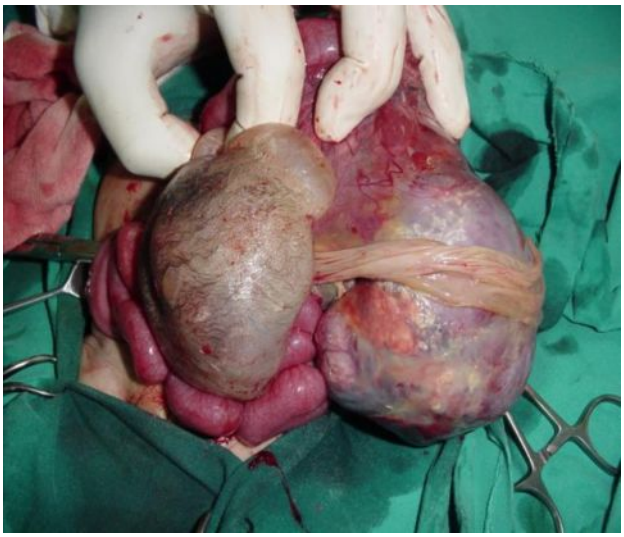

Fig. 3: Fetus In Fetu

Fig. 4: Teratopagus Twin

Fig. 5-A: Thoraco-Omphalopagus Twin

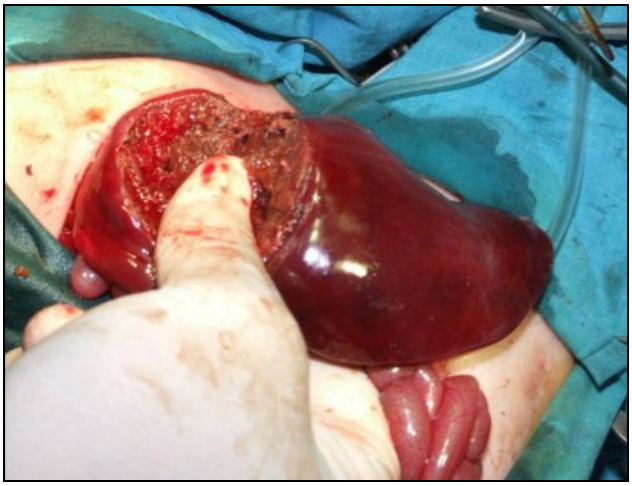

Fig. 5-B: Separation with Finger Fracture Technique

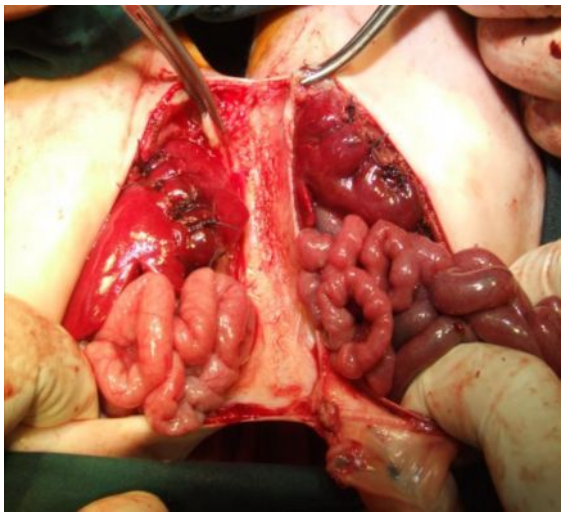

Fig. 5-C The Last Area Before Complete Separation

Fig. 5-D: Ten Days After Separation

DISCUSSION

Conjoined twins have fascinated people and challenged surgeons for years, their management remains exciting and challenging [15]. CT has high mortality. In utero death rate was 28% and soon after birth 54% with only 8% survival rate [16].

Operation timing and separation methods vary in types of anomalies and the conditions of the fused organs, associated malformations and general conditions of the infants [17].

Spitz and Kiely [5, 18] have indicated that elective separation of CT is safe between 2 and 4 months of age, however emergency situations that may force separation immediately include the presence of a stillborn twin, intestinal obstruction, rupture of an omphalocele, heart failure, obstructive uropathy or respiratory failure [17].

Like the case presented by Hilfiker et al. [19] we have postponed our case of thoraco-omphalopagus CT till one month of age to help maturation of the babies organs and to help weight gain with no concern about infection but Hilfiker et al., [19] used rapid tissue expansion technique to provide skin coverage which was not needed in our case, where the defect was closed by mobilization of skin flaps plus making lateral release incisions in one of the twins. On the other hand EL-Gohary [20], through his series in UAE operated a similar case of omphalopagus twins at about 52 days of life.

Rapid separation of CT may be needed in the first days of life, as in our case of epigastric heteropagus twins and the case of oral parasitic teratopagus twin where the parasitic twin caused respiratory embarrassment for the autosite, the same have been performed for the pygopagus twins where the malformed twin died, which necessitated rapid separation at the 3rd day of life, similarly votteler and lipsky [21] during the period from 1978 to 2000 performed six separations emergently because of death or impending death of their respective twins, mean while they performed an elective separation of one of the twins at 3 weeks of age.

The incidence of epigastric heteropagus twins is very rare with only 25 reported cases, male predominance is seen among the reported cases. [7, 21-25] .I think that our case is the twenty six reported one. The condition may result from ischemic atrophy of one part early in the gestational life [22].

Similar to our case of oral teratopagus twin, EL-Gohary [20] reported a more organized parasitic twin projecting from the mouth of the autosite but had an element of embryonal carcinoma causing recurrence in the submandibular area that was successfully removed.

In omphalopagus CT, some degree of hepatic fusion is almost inevitable and a conjoined small intestine is reported in up to 50% of cases. Conjunction of the biliary tree was also reported in 25% of cases [23]. In our case of omphalopagus CT, there was a single liver with separate biliary systems. The liver was divided to its anatomical right and left lobes, each having separate extrahepatic biliary systems, there was no small intestinal connections. This in reverse to the case presented by Spitz et al [24] where there was a liver for each baby and joined by small bridge of tissues, but the small intestine was common for both.

In the cases presented as fetus in fetu, the parasite embodied in the autosite, usually within the cranial, thoracic or abdominal cavities of the autosite which may occur due to anastomosis of vitillene circulation.

CONCLUSION

- Careful planning and experience are important factors in dealing with CT.
- Parasitic twins and twins with sever anomalies incompatible life in one of them are considered to be one person.
- Timing separation and separation plan should be individualized according to the need of emergent separation and the degree of organ fusion.
- The lesser the fusion of organ systems, the greater the ultimate functionality and survival

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